A CASE OF MULTIPLE FIBROMATA CONFINED TO THE INTERNAL PLANTAR NERVE.[1]

By Wm. J. Taylor, M.D., and Wm. G. Spiller, M.D.

REMARKS BY DR. WM. J. TAYLOR.

The case which I now report is to me unique and many of its features are of such interest that it seems worthy to be placed on record. I first saw this patient, then a young woman of twenty-seven and a mill-hand, in the autumn of 1888. Her mother was living and well, her father had died from accident, and she had five brothers and sisters all living and well. She had had smallpox when a child, but no other serious disease, and no specific history, nor has she ever had rheumatism. When about sixteen years of age she sprained her left ankle and since then it had always been weak and had a tendency to turn inward. In 1881, or seven years before she was seen by me, she began to suffer from pain and soreness in the sole of the left foot and inner side of the ankle. At first this was noticed only upon pressure, but gradually it grew worse until she could hardly stand upon the foot or sit with the foot in a dependent position. The pain at that time was not so severe on walking, but the foot was only free from pain when she was lying down. For two years before she was first seen, or in 1886, her work had been such as to require a good deal of standing, as she was working in a mill, and this seemed to aggravate the condition. For the past few months this pain had become more localized and was confined to spots along the inner side of the foot and ball of the great toe. After an attack of unusually severe pain the foot would swell and become red, and it would also swell at any time if she sat with the foot in a dependent position.

At this time, October 29, 1888, many small masses could be felt along the inner side and sole of the foot—these masses varied in size from that of a hickory nut to that of a small pea, and followed the distribution of the internal plantar nerve. They were movable, hard, intensely painful to the slightest touch, and some of them seemed to be immediately beneath the skin, while two small masses could be felt on the ball of the great toe; in all about ten small masses could be distinctly outlined. At times they became red and inflamed. They were so exquisitely sensitive that the slightest touch would give her agonizing pain—simply drawing the finger lightly across the skin of the sole of the foot would cause her almost to fall off the chair upon which she was sitting. The whole foot and particularly the sole of the foot sweated so, and particularly after any manipulation, that the sweat would stand out in beads all over the skin, and at times dropped to the floor.

---

[1] Read before the Philadelphia Neurological Society, Jan. 27, 1903.

On November 1st, 1888, she was operated upon by Dr. William Hunt at the Orthopedic Hospital, where as Assistant Surgeon, I had the opportunity of helping him. Thirteen small tumors were removed from along the line of the distribution of the internal plantar nerve. These masses varied in size from that of a hickory nut down to a very small pea. They were shelled out from beneath the skin with the greatest ease, having very slight attachments. They were quite firm and hard in consistency and contained no fluid when cut open, but showed simply a white and glistening surface; one or two of the larger ones seemed to be composed of three or four small ones together. Following this operation she had quite a period of comfort, but at no time was she entirely free from pain.

In 1890 she was readmitted to the hospital under my care, stating that she had been much better for about a year after the first operation, but the pain had returned and was quite as severe as ever. I now operated and removed two tumors, one at the instep and the other at the ball of the great toe. Following this she had a period of comfort, but was readmitted in 1897 when I again operated and removed six tumors of the same general character and from the same region, one the size of a marble from in front of the tendo Achillis, and above the level of the ankle joint; again she was relieved. These tumors were pronounced to be pseudo-neuromata by Dr. Guiteras. All manner of medication was tried—iodine, tonics, iodide of potassium, different types of apparatus, bandages—in fact everything possible that could be suggested, was experimented with. During this time the pain recurred, varying in intensity, but of such a character that she was obliged to give up her occupation, that of a mill-hand, as she could not stand on her feet. There seemed to be a reproduction of the small tumors which increased in number and grew in size until June, 1902, when she was again admitted to the hospital, stating that the pain was of such a character that she could not possibly stand it any longer. The foot was now intensely sensitive —four or five tumors could be distinctly outlined—the slightest touch even to the skin over one of these tumors caused intense pain and profuse sweating of the foot.

On June 13th, 1902, I operated, making a long incision over the posterior tibial nerve just above the internal malleolus, and extending it down along the distribution of the internal plantar nerve, quite eight inches in length. Five small tumors were found along the course of this nerve and were removed; one large tumor was found deep down in the sole of the foot beneath the dense fascia, and it was only distinctly located by its extreme sensitiveness. Even when she was under the influence of ether pressure would cause her to wince. This tumor was especially large, about the size of a small walnut, and was attached to the internal plantar nerve at several points by rather long bands apparently of nerve tissue. The internal plantar nerve was found greatly enlarged, larger than an ordinary lead pencil. In view of her long-standing pain, I exposed and removed about five inches of this greatly swollen nerve, to which were attached several of these tumors. The wound healed without the slightest difficulty and all sensation was gone. For the first time in these many years (14) she was free from pain and although all sensation over a certain area was absent, her joy was very great. Her general health improved to a wonderful degree, and since that time she has been perfectly well and able to resume her work.

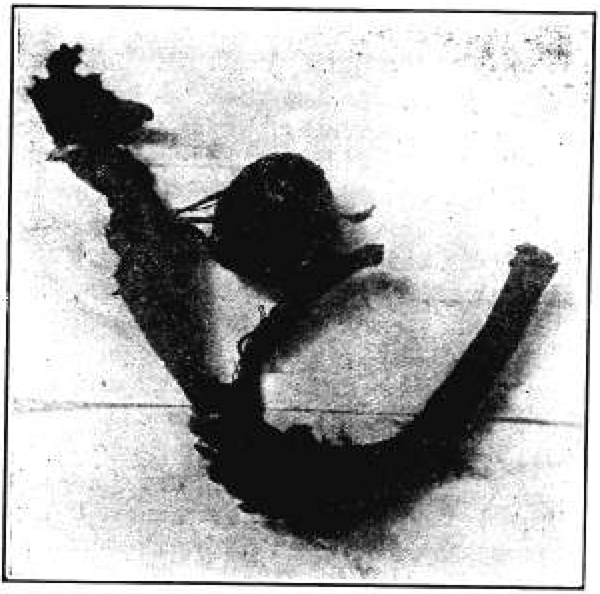
As I have stated, this case in my experience is absolutely unique, and Dr. Wm. G. Spiller, who has been kind enough to make microscopical examinations of the specimens, will report the neuropathological conditions.

November 29th, 1902, I examined the condition of the foot and found to be remarkably good. There is no pain whatever on walking, nor standing, nor has she at any time pain. The foot has been weak until very lately, but now it seems to have almost entirely regained its strength. Sensation is perfectly good in the skin of the sole of the foot, and there is tenderness at only one place in the whole line of the scar, and that is directly at the upper portion about on a level with the cut end of the nerve behind and above the malleolus. I imagine this sensitiveness must be simply at the end of the nerve, for I was not able to go entirely above the point of swelling in the nerve itself. The foot no longer sweats at all. She tells me that she is perfectly well, has great comfort and is able to work steadily.

REMARKS BY DR. WM. G. SPILLER.

The report of a case of multiple fibromata confined to the ulnar nerve was published by Dr. W. W. Keen and myself in May, 1900. In that paper reference was made to the case of Dr. W. J. Taylor, now described at length. In view of the fact that Garrè, in speaking of his case, remarked that the presence of multiple fibromata in the skin of the sole of the foot was very extraordinary, and that v. Recklinghausen emphasized the immunity of this part and of the palm of the hand, it seems strange that in the two cases in which I have been concerned the hand was affected in one and the sole of the foot in the other. In one case the tumors were confined to the ulnar nerve, and in the other to the internal plantar. In Dr. Taylor's case a sprain of the left foot preceded the development of the symptoms of fibromata about four years, and the ankle after the injury remained weak. This accident may have been the immediate cause of the proliferation of the fibrous tissue in the left internal plantar nerve in a person predisposed to such proliferation. Dr. Keen's patient was a laborer, and trauma possibly aided in the formation of nerve tumors in the palm of the left hand. Trauma alone does not explain this singular process, or else multiple fibromata confined to a single nerve would be common, instead of being a very rare finding. Indeed, they seem to be more uncommon than the generalized neurofibromatosis. In the paper with Dr. Keen I referred also to a case of tumors confined to one nerve, reported by J. K. Mitchell and one by Bowlby.

In some cases heredity plays an important rôle, and members of different generations in the same family have exhibited neurofibromatosis. (Menke, Herczel, Bruns, Czerny.) It is impossible to believe that the process is purely an acquired one, and even in the cases in which the tumors have been confined to one nerve it is probable that the tendency to proliferation exists in other nerves, and that the proliferation may be called forth by trauma, or possibly by disease of the blood vessels. In the specimens sent to me by Dr. Taylor for examination the blood vessels removed with the nerve and tumors are greatly diseased, and the walls of the arteries are much thickened; some vessels indeed are thrombotic and without any lumen. This interference with the nutrition of the nerve may have aided in the production of fibrous tissue within the nerve bundles. The tumors are fibromata and contain only such nerve fibers as have not become degenerated by pressure; they therefore are not true neuromata in the sense that proliferation of true nerve tissue has occurred. In amputation neuromata there is a formation of young nerve fibers with the proliferation of fibrous tissue, so that these are truly neurofibromata, but in the specimens I have examined from Dr. Keen's patient and in those from Dr. Taylor's patient the fibrous tissue alone is proliferated.



A tumor dependent from the nerve trunk by several bands of fibers.

In one of the small tumors removed by Dr. Taylor which I have cut lengthwise and examined microscopically, medullated nerve fibers may be seen entering the tumor at one end, and then after entering separating and passing to the periphery of the tumor, and gradually disappearing from view, so that where they become widely separated from one another they can not be traced at all. As in Dr. Keen's case the proliferation has probably begun in the endoneurium of the nerve, and therefore the tumors are somewhat elongated, with their long axes parallel to the nerve. As the perineurium becomes thickened it offers some resistance to further growth of the tumor, and the proliferation of the fibrous tissue occurs more rapidly where resistance is less, viz., at the two ends of the tumor.

The tumors removed by Dr. Taylor, with one exception, are small, as were those removed by Dr. Keen from his patient.

It is easy to discover the cause of the intense pain in these cases. The nerve fibers are pressed upon and irritated by the proliferating connective tissue, and although many nerve fibers degenerate and thus lose the power of conducting painful stimuli, the process is progressive, and new nerve fibers are attacked as those previously diseased disappear.

One tumor, the size of a small walnut, removed by Dr. Taylor, is of peculiar interest, because it is dependent from the large nerve trunk by several thin bands of fibers. These seem on inspection to be bands of connective tissue, but one of these bands cut in microscopical sections consists of nerve fibers embedded in and surrounded by much fibrous tissue. This tumor has evidently grown from several small nerves, or else it has grown from one nerve and surrounded and embedded others within it.

The long duration of the process in Dr. Taylor's case and the repeated operations deserve mention, because removal of these fibrous proliferations of nerves has been regarded by some writers as liable to cause a change from fibrous into sarcomatous tissue. This has not occurred in Dr. Taylor's case.

I have discussed the subject of neurofibromatosis at length in the paper published in collaboration with Dr. Keen in May, 1900, and shortly after the appearance of this paper the excellent monograph on the same subject by Alexis Thomson was published. It is unnecessary therefore to say anything further concerning the specimens from Dr. Taylor's case.